WHAT IS CLAIMED IS:

1. A human α1,2-mannosidase enzyme for specifically converting $Man_9GlcNAc$ to $Man_8GlcNAc$ isomer B in degradation mechanism of misfolded proteins, wherein said enzyme has the characteristics of an enzyme encoded by a cDNA sequence set forth in Fig. 1.

2. An agonist or antagonist of the α1,2-mannosidase enzyme of claim 1 for activating or inhibiting said enzyme.

3. The agonist or antagonist of claim 2, wherein said activating or inhibiting is for a transient period of time.

4. An antagonist of claim 2, wherein said inhibiting is for a transient period of time, thereby preventing degradation of misfolded glycoproteins.

5. A method for the treatment of a genetic disease causing a misfolding of proteins in a patient, which comprises administering an antagonist of α1,2-mannosidase enzyme of claim 1 for transiently inhibiting said enzyme, thereby preventing degradation of misfolded glycoproteins.

6. The method of claim 5, wherein said genetic disease is selected from the group consisting of cystic fibrosis and emphysema.

7. The method of claim 6, wherein for cystic fibrosis said misfolded protein is cystic fibrosis transmembrane conductance regulator (CFTR).

8. The method of claim 6, wherein for emphysema said misfolded protein is alpha1 antitrypsin.

9. The use of a mutant α1,2-mannosidase to produce altered recombinant glycoproteins with improved uptake.

10. The use of claim 9, wherein said improved uptake is to treat genetic diseases characterized by formation of misfolded glycoproteins resulting in their degradation and/or improper localization.